Letter to the Editor: "Bad-Risk Childhood Pleuropulmonary Blastoma: Does Chemoradiotherapy Help?"

We have read with interest the report and review by Schmaltz et al. [1] on the management of a case of childhood pleuropulmonary blastoma. According to the article, both conventional chemotherapy and ultimately megatherapy were employed. Here we contribute our own experience with a similar case, a 4-year-old girl, who was treated with repetitive chemotherapy regimens as well as radiotherapy and who survived for 22 months following diagnosis.

The girl was admitted to our department with a 1-month history of cough, temperature, and gradually deteriorating dyspnea refractory to treatment.

On chest X-ray a large right medium and lower pulmonary field shadow was revealed, which on CT exhibited the features of an inhomogeneous mass with pleural effusion.

Needle biopsy revealed a non-specified malignancy consisting of spindle-shaped neoplastic cells.

Ensuing right thoracotomy resulted in gross excision of a multilobal tumor measuring 8 × 9 × 12 cm in size, adjacent to the right lower lobe and the adjoining pleura which was found infiltrated.

Histology proved to be that of a childhood-type biphasic pleuropulmonary blastoma [2–4], a diagnosis that was confirmed after having been independently reviewed (Leake and Pritchard, 1993). Staging revealed no distant metastatic spread. Due to microscopic residual disease we complemented surgery with ''sandwich'' chemoradiotherapy. She received 5 cycles of IVAD (ifos 9 g/m^2, vincristine 1.5 mg/m^2, doxorubicin 60 mg/m^2), followed by irradiation, a total of 40 Gy in 20 fractions and then a further 2 IVAD and 3 IVA (ifos, vincristine, actinomycin D 1.5 mg/m^2) courses. Following chemoradiotherapy the patient remained disease-free for 6 months (14 months post-diagnosis). She subsequently relapsed locally. On restaging both hepatic and bone metastases became apparent.

Relapse was treated with chemotherapy (4 cycles of cisplatin 150 mg/m^2, VP-16 300 mg/m^2), which maintained the disease stable for another 5 months. She also received concomitant palliative irradiation to her painful bone metastases. As the disease subsequently progressed, she presented with pronounced hypercalcemia successfully treated with sodium pamidronate administration.

The patient died 3 months later, having survived a total of 22 months from diagnosis.

In spite of the highly malignant potential of the disease and its dismal prognosis, we succeeded in maintaining our patient disease-free for 6 months by intensive chemotherapy coupled with irradiation without utilizing megatherapy. It is our impression that intensive chemoradiotherapy could improve treatment efficacy of bad-risk childhood pleuropulmonary blastoma [5,6].

We have recently also come across the paper by Lobo-Sanahuja et al. [7] on the management of childhood advanced pulmonary blastoma by surgery followed by conventional intensive chemotherapy. We believe that the addition of radiotherapy to intensive chemotherapy may further improve event-free survival and quality of life.

K. Pistevou-Gompaki, PhD
Department of Radiotherapy
Theagenion Hospital

N. Gompakis, PhD
First Department of Paediatrics
University of Thessaloniki

M. Trachana, PhD
Department of Paediatric Oncology

E. Kostopoulou, PhD
Department of Histopathology

D.E. Koliouskas, PhD
Department of Paediatric Oncology
Ippokration Hospital
54642 Thessaloniki, Macedonia, Greece

REFERENCES

1. Schmaltz C, Sauter S, Opitz O, Harms D, Kremens B, Lohner M, Metz K, Brandis M, Niemeyer C: Pleuro-pulmonary blastoma: A case report and review of the literature. Med Pediatr Oncol 25: 479–484, 1995.
2. Manivel JC, Priest JR, Watterson J, Steiner M, Woods WG, Wick MR, Dehner LP: Pulmonary blastoma: The so-called pulmonary blastoma of childhood. Cancer 62:1516–1526, 1988.
3. Delahunt B, Thomson KJ, Ferguson AF, Neale TJ, Meffan PJ, Nacey JN: Familial cystic nephroma and pleuropulmonary blastoma. Cancer 71:1338–1342, 1993.
4. López-Andreu JA, Ferris J, Esquembre C, Verdeguer A, Gómez J, Castel V: Familial cystic nephroma and pleuropulmonary blastoma (letter). Cancer 72:2792, 1993.
5. Koss MN, Hochholzer L, O'Leary T: Pulmonary blastomas. Cancer 67:2368–2381, 1991.
6. Ohtomo K, Araki T, Yashiro N, Iio M: Pulmonary blastoma in children. Radiology 173:77–79, 1989.
7. Lobo-Sanahuja S, Garcia I, Santa Maria S, Barrantes JC: Case report: Pulmonary blastoma in children—Response to chemotherapy. Med Pediatr Oncol 26:196–200, 1996.